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RETROPERITONEAL GANGLIONEUROMA: A CASE REPORT

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A primary extra-adrenal retroperitoneal ganglioneuroma was found incidentally in a 17-year-old boy during a series of examinations for hematopyuria. Computed tomography and magnetic resonance imaging revealed a homogenous mass located between the right proximal ureter and the inferior vena cava just below the right kidney. Subtotal resection of the tumor was performed through a transabdominal approach. The resected specimen measured 8.5×6×5.5 cm and weighed 70 g. Histological examination showed that the lesion was an extra-adrenal retroperitoneal ganglioneuroma composed of mature ganglion cells and nerve fibers without any malignant features. The patient has shown no signs of recurrence in the 4 months after surgery. We provide a review of the Japanese literature on extra-adrenal retroperitoneal ganglioneuroma and also discuss the clinical features of this tumor.

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Key words: Ganglioneuroma, Retroperitoneal tumor

INTRODUCTION

Ganglioneuroma is a relatively rare benign neoplasm which originates in the sympathetic nervous system. The most common site involved is the posterior mediastinum, but this tumor may also occur in the retroperitoneal space, the pelvic cavity or the adrenal gland[1] Ganglioneuroma apparently represents the most mature form of the neuroblastoma-like tumors[2]

These are slow-growing tumors and they are often asymptomatic. However, improvements in imaging have led to an increase in the detection of these tumors in recent years. We herein report a case of extra-adrenal retroperitoneal ganglioneuroma that was found incidentally during the investigation of hematopyuria and review the Japanese reports on this tumor.

CASE REPORT

A 17-year-old boy presented to our hospital with macrohematuria. Urinalysis showed hematopyuria and Escherichia coli (E. coli) was identified by culture of a urine specimen. The infection was rapidly cleared by antibiotic therapy. Excretory urography revealed lateral deviation of the right proximal ureter, but no hydronephrosis was present (Fig. 1). Computed tomography and magnetic resonance imaging (Fig. 2) revealed a mass in the retroperitoneal space adjacent to the right proximal ureter. The lesion appeared to be encapsulated. Angiography and enhanced computed tomography demonstrated that it was hypovascular.

Based on these findings, the lesion was thought to be a tumor of the nervous system. Excision of the lesion was performed via the transabdominal approach. At the operation, a tumor was found lying between the right proximal ureter and the inferior vena cava. Both the ureter and the vena cava were not invaded by the tumor, but because of extensive adhesion to the vertebrae complete resection of the mass was impossible. The resected tumor measured 8.5×6×5.5 cm and weighed 70 g.

Histological examination showed that the lesion was composed of nerve fibers and mature ganglion cells (Fig. 3). In some areas, the ganglion cells were aggregated or proliferating in a neoplastic fashion and Schwann cells were frequently seen. However, no evidence of malingancy could be found in any specimen examined.

Thus, the histological diagnosis was retroperitoneal ganglioneuroma.

DISCUSSION

Ganglioneuroma is a benign neoplasm which originates from the sympathetic nervous system and is composed of mature ganglion cells and nerve fibers. This tumor was first recognized as an independent entity by Loretz in 1870[3]. The posterior mediatinum is the most common site of origin, but this tumor may also develop in the retroperitoneum or the adrenal gland. Ganglioneuroma is reported to constitute 0.72% of all retroperitoneal tumors[4].

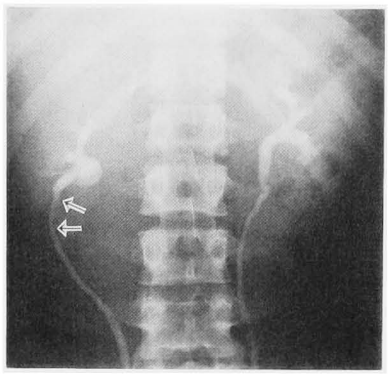

Fig. 1. Drip infusion pyelogram shows lateral deviation of the right proximal ureter (arrow), suggesting the existence of a retroperitoneal mass.

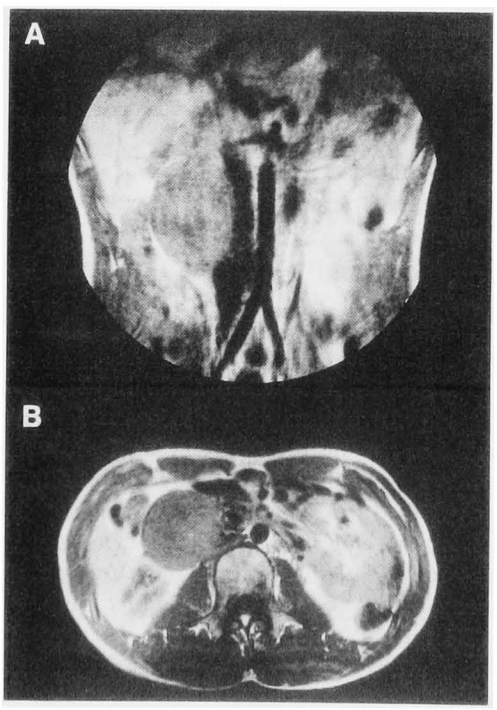

Fig. 2. (A) Coronal magnetic resonance image (MRI) demonstrates a retroperitoneal tumor adjacent to the inferior vena cava. (B) On transverse MRI, the tumor appears to be encapsulated but also seems to adhere to the vertebrae.

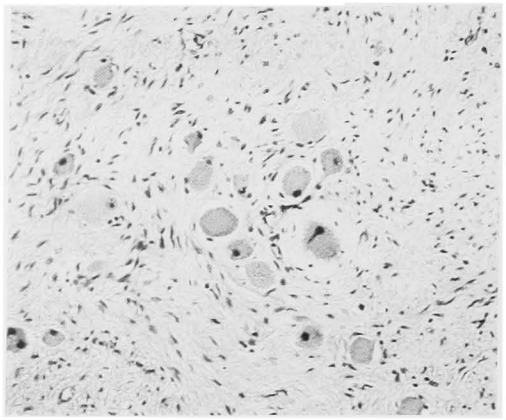

Fig. 3. Microscopic examination showed that the tumor was composed of nerve fibers and mature ganglion cells (H.E. ×100).

Carpenter and Kernohan reported that this tumor has a slight female predominance[5]. However, a review of the Japanese literature shows that ganglioneuroma occurs with almost equal frequency in both sexes in this country. The age distribution of the Japanese cases is in agreement with that generally reported showing a preponderance in the young (60% of the Japanese patients were less than 30 years old and the mean age was 25.3 years).

Ganglioneuroma is a slow-growing tumor that often becomes quite large before detection. Among the Japanese patients with ganglioneuroma, only 40% had symptoms referable to the tumor itself.

Along with various dramatic developments in imaging diagnosis, the detection of this neoplasm during the evaluation of other diseases has increased in recent years.

Hormonally active ganglioneuromas, which are associated with increased levels of urinary cathecholamines, homovanillic acid, or vanillyl mandelic acid, are predom-

inantly found in pediatric patients. The Japanese literature includes reports on 8 cases of hormonally active extra-adrenal retroperitoneal ganglioneuroma. All these tumors occurred in patients aged younger than 6 years.

Even since Cushing and Wolbach reported on the apparent maturation of a neuroblastoma to a ganglioneuroma in 1927[6], tumors of neural crest origin have been thought to occasionally undergo transformation from malignant neuroblastoma to benign ganglioneuroma. However, only a few such cases have been reported in Japan[7].

This tumor should be surgically excised as completely as possible in order to distinguish it from neural crest malignancy. Even if excision is incomplete, the prognosis is generally reported to be quite favorable. If the tumor shows no malignant features histologically, no additional management is needed apart from periodical postoperative review for the early detection of recurrence.

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和文抄録

後腹膜神経節細胞腫の1例

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野々村祝夫，菅野　展史，妹尾　博行
赤井　秀行，武本　征人

後腹膜原発の副腎外神経節細胞腫を経験したので報告する．腫瘍は17歳男性の血膿尿精査中に発見された．CT および MRI 検査にて右腎直下に，近位部尿管と下大静脈の間に位置する内部均一像を示す腫瘍を認めた．内分泌学的検査では異常は認められなかった．経腹的に切除した腫瘍は 8.5×6×5.5 cm 大，重量 70 g であった．病理組織学的には，腫瘍は神経節細胞と神経線維からなる神経節細胞腫であり，悪性所見は認められなかった．副腎外に発生した後腹膜神経節細胞腫の本邦例における文献的考察を加える．

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